

Typical absence seizures in adults: clinical, EEG, video-EEG findings and diagnostic/syndromic considerations

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Abstract

Eighteen women and five men had typical absences. These included 10% of a consecutive hospital series of 200 adult patients with epileptic disorders. The absences began between the ages of seven and 46 years and varied in type and severity. Twenty patients also had generalised tonic-clonic seizures, ranging in frequency from one in a lifetime to one per month. Myoclonic jerks of the limbs occurred in 11 patients but were not associated with the absence attacks. Eyelid myoclonus consistently occurred with absence attacks in four patients and perioral myoclonus in two patients. Absence status occurred in five patients. Absence seizures were frequently unrecognised or misdiagnosed as complex partial seizures. Satisfactory control was achieved with sodium valproate. Electroencephalography, particularly video-electroencephalography, was invaluable in the diagnosis, but focal abnormalities in seven patients might have been erroneously interpreted as indicating partial seizures. This series showed that clinical and EEG manifestations are often syndrome-related and that there are more epileptic syndromes with typical absences than those presently recognised.

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Though often considered rare in adults, typical absences with onset in childhood and puberty persist into adult life in 7-81% of cases.¹⁻¹⁹ Loss or severe impairment of consciousness is considered to be the clinical hallmark of absences.^{20, 21} EEG manifestations of typical absences, however, are frequently associated with mild, inconspicuous cognitive impairment.²²⁻²⁴ Furthermore, typical absences have been studied in a uniform fashion^{1-19, 25, 26} despite existing evidence that their clinical and EEG features are syndrome-related.²⁴ Thus prognosis has been based on a symptom (typical absences)²⁰ rather than a syndrome/disease diagnosis.²¹

This is the first clinical, EEG and video-EEG study of typical absence seizures and syndromes in adults.

Methods and patients

Twenty patients with typical absences, as defined by the Commission on the Classifica-

tion and Terminology of the International League Against Epilepsy (ILAE),²⁰ were identified from the first 200 consecutive clinical referrals between April 1989 and April 1991 to a new epilepsy clinic in St Thomas' Hospital. Three additional patients, referred for the purpose of this study, were included.

Patients were referred from accident and emergency departments, general practitioners, physicians and other neurologists within St Thomas' Hospital and the south-east Thames region. Eighteen of these 23 patients had been seen by at least one consultant neurologist and they all had had one or more EEGs.

Clinical and EEG evaluation of all patients was made by one of us (CPP). Diagnosis was based on the proposal of ILAE,²¹ when not possible, a seizure-symptom categorisation²⁰ and a differential syndromic diagnosis was made. Early clinical records and EEG reports were obtained and scrutinised.

Routine EEG and video-EEG (34 channel-Telefactor) recordings were carried out using previously reported techniques.^{23, 24} Patients were asked to count their breaths out loud during hyperventilation, which allowed us to evaluate impairment of consciousness and speech during generalised spike-wave discharges.^{23, 24} Twelve patients were studied with video-EEG and 83 recorded absences were analysed.

Results

Prevalence, sex, age

Ten per cent of the 200 unselected patients had typical absences. There was a threefold female predominance of the total group (18 women, 5 men). All patients were over 20 years of age with normal neurological and mental status. Table 1 shows the age at referral, onset of seizures and duration of the disease.

Absences

All patients had absences documented with EEG or video-EEG or both. The age at onset of absences is shown in table 1. Late-onset absences, occurring after the second decade of life, were reported by six patients. Onset of absences could not be estimated by six patients and another one (case 23) remained unaware of them despite video-EEG confirmation.

The absences were subjectively perceived by the patients in a stereotyped way and were described as brief "flashes of blackouts", "like in a daze", "getting into a trance" (table 2). The degree of impairment of consciousness from mild "momentary lack of concentration"

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Case No	Sex	Age	Onset (years) of			Frequency of		Classification
			Absences	Myoclonic jerks	GTCS	GTCS	Absences	
1	F	22	Not known	13		None	Occasional	JME
2	M	29	Not known	13	24	3/life	Frequent	JME
3	F	25	Not known	16	20	1/life	Occasional	JME
4	F	40	7	10	12	1/month	2/day	JME
5	F	25	Not known	13	13	4/life	Occasional	JME
6	F	41	Not known	16	22	4/year	3/day	JME
7	F	26	11	13	14	Not available	Occasional, evoked	JME
8	F	42	18		35	1/life	3–4/day	Eye-lid myoclonia with absences
9	M	43	30	35	9	4/year	1–5/day	Eye-lid myoclonia with absences
10	F	38	20		11	1–3/year	6/month	Eye-lid myoclonia with absences
11	M	26	12		12	1/month	Several/day	JAE
12	F	44	11		12	1–2/year	1–2/year	JAE
13	M	38	12			None	1–2/month	JAE
14	F	39	8			None	Many/day	JAE
15	F	46	46		46	2/life	2–4/life	Absence status
16	F	34	24		12	TV induced	Frequent, evoked	Photopattern sensitive epilepsy
17	F	30	28		28	2/life	1–2/month	Symptomatic absence epilepsy
18	F	35	15	18	25	1–2/year	Infrequent	Atypical JME? JAE?
19	M	60	Not known	35	9	1–2/year	2/month	Atypical JME? JAE?
20	F	21	7		7	1/year	5–6/day	Perioral myoclonus with absences
21	F	27	13		22	3/Life	Frequent	Perioral myoclonus with absences
22	F	67	30		30	1/year	1/month	Late-onset absences/absence status?
23	F	27	Unaware			1–2/life	Unaware	Late-onset absences with GTCS?
Mean	35.8	18.3	18.2	19.1				
SD	11.3	10.5	8.7	10.1				

GTCS = generalised tonic-clonic seizures; JME = juvenile myoclonic epilepsy; JAE = juvenile absence epilepsy.

Case no	Description
1	Like in a distance, mind becomes blank, thoughts jumbled.
2	Flashes of loss of concentration, tiny/little blackouts.
3	Momentary lack of concentration.
4	Momentary lack of concentration, blanks, dream, deja-vu, strange and terrifying feelings.
5	Brief episodes of absent mindedness.
6	Blank, going to a distance, lack of awareness, you are there and not there.
7	Momentarily loss of concentration.
8	Loss of concentration,unresponsive.
9	Feels away from things and unreal.
10	Gets into a trance, like day-dreaming, not responding to commands.
11	Loss of concentration-like being hit on the back of the head—like in a daze.
12	Like in a trance.
13	Discontinuation of what one thinks or says, a very quick black-out.
14	Blank spells, unresponsive.
15	Becomes slow, not herself.
16	Momentary loss of contact, freeze in front of the kitchen carpet.
17	Mentally as if I slipped a cog.
18	The world around me seems to have moved without me, lost the thread of what they had said.
19	In and out of a state of consciousness, disorientated behaviour, some jerking.
20	Blank episodes, blurring as if in a dream, unresponsive.
21	Vacancies, distracted, unable to understand basic commands.
22	Strange episodes of inappropriate behaviour, confused.
23	Momentary loss of track of things.

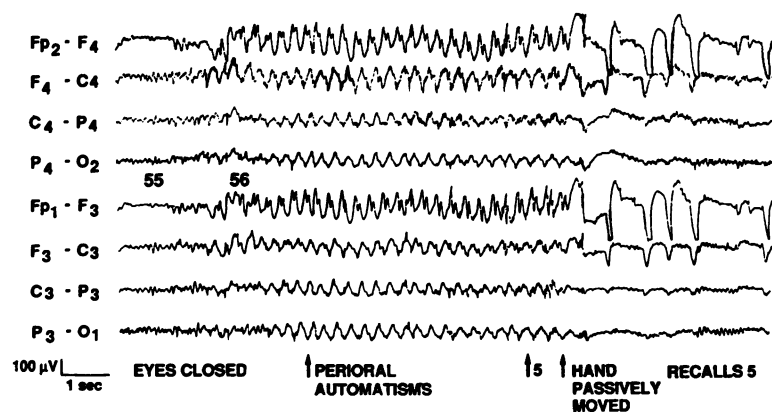


Figure 1 Video-EEG of a 39 year old woman (case 14) who had severe and frequent absences since age 8 years without myoclonic jerks or generalised tonic-clonic seizures. The absence seizure illustrated was recorded during breath-counting in hyperventilation. Her eyes remained closed, she stopped overbreathing and counting but, with some hesitation, pronounced correctly the subsequent number (56) after the onset of the discharge. Ictal oral automatisms were observed. She could remember the number five spoken to her immediately before the end of the ictus. The seizure was terminated by passively moving her hand.

De-realisation and fear were experienced by 2 patients (cases 4 and 9). Automatisms were never reported by witnesses or by patients but were recognised in video-EEG studies in one patient (fig 1, case 14). Facial myoclonus localised in the eyelids (fig 2, cases 8–10, 17) or perioral muscles (cases 20–21), of which the patients were unaware, was demonstrated during the absences with video-EEG or described by witnesses. Severity and frequency of absences varied from causing little disturbance to imposing a serious social and professional handicap to the patient. Absence status was documented with EEG or video-EEG, or both, in four patients (cases 15, 19, 21, 22, fig 3) and suspected in a fifth (case 12). Episodes comprised protracted periods (5–12 hours) of mild to moderate confusion. In four cases absence status was unrecognised by medical and nursing personnel for many years.

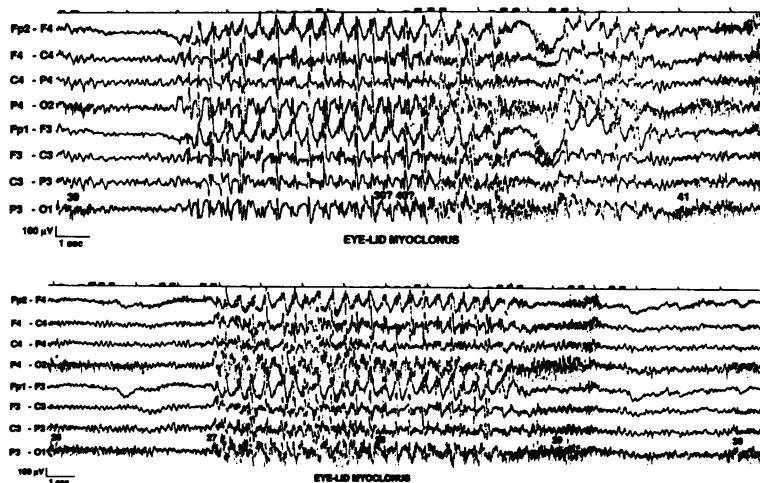


Figure 2 EEG of a 43 year old patient (case 9) who had onset of GTCS at age 10 years. He became aware of absences at age 30 years. Eyelid myoclonus was consistently associated with the ictus. Discontinuation of breath counting was observed in one (upper trace) but not another (lower trace) of the multiple-spike wave generalised discharges. These discharges were consistently associated with eyelid myoclonus but the severity of cognitive impairment showed a considerable variation, probably depending on his pre-ictal level of alertness.

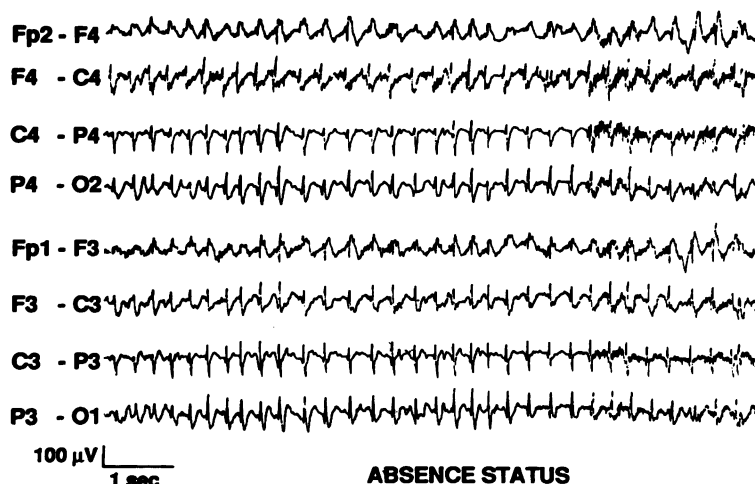


Figure 3 Absence status recorded by video-EEG of a 67 year old woman (case 22) who was treated with primidone and sulthiame for 37 years. Her GTCS were preceded by a state of mild confusion, including "strange episodes of inappropriate behaviour like [sic] putting on her trousers over her pyjamas" which were interpreted as complex partial seizures. This video-EEG recording, seven hours after onset of symptoms, demonstrated absence status. Clinically she was slow in her answers and moderately confused. Status was terminated with diazepam iv and sodium valproate iv. Note fast 3–4 Hz spike-slow wave activity which was continuous.

Myoclonic jerks

A clear distinction between limb myoclonic jerks and localised facial (eyelid or perioral) myoclonus was apparent. Limb myoclonus did not occur during the absence ictus, except during myoclonic absence status. Conversely, facial myoclonus alone (eyelid in four patients and perioral in two) was always associated with the absence ictus. Myoclonic jerks of the limbs occurred in 11 patients: on awakening in seven (cases 1–7), random, diurnal and usually mild in three (cases 9, 18, 19) and induced by television in one (case 10).

Generalised tonic-clonic seizures

Generalised tonic-clonic seizures (GTCS) were reported by 20 patients (table 1) and

varied in frequency from one in a lifetime (for example, case 8) to several per month (case 4). The age at onset ranged from 7–46 years (mean 19.1 (SD 10.1)). Although absences occurred first, it was often a GTCS that precipitated medical attention.

EEG and video-EEG studies

Absences were recorded in all patients. Previous clinical records and EEG reports revealed that some patients had spike-slow wave discharges long before typical absences were clinically recognised, such as in case 18.

The duration of the discharges varied from 1–2 to 20 s but lasted for hours in absence status. Discharges were generalised, of higher amplitude in the anterior regions, with dominant intradischarge spike-wave frequency of 3–4 Hz (complexes of 2–6 Hz were also seen). In some patients, such as cases 13 and 14 (fig 1), the discharges consisted of repetitive spike-slow wave complexes with steady fall in frequency from the onset to the termination of the paroxysms. In others (cases 1–10 mainly, figs 2, 4) there were frequent multiple spike components and discharge fragmentations.^{23 24}

The paroxysms of patients 1–7 were often irregular, with a variation of the intradischarge frequency (fig 4).

The discharges were associated with impairment of consciousness which greatly varied in severity. Some patients had clinical manifestations during spike-wave paroxysms as short as 1.5–2.5 s (fig 5). In others, or even the same patient, however, there were no discernible mental changes in much longer EEG discharges (fig 2), although in one patient impairment of consciousness was often proportional to the length of the paroxysm. Automatisms were recorded in one, and facial myoclonus in four patients. A discharge often could be interrupted by auditory or somatosensory stimuli or both.

Focal paroxysmal abnormalities were recorded in seven patients. These were short transients of localised slow, sharp waves or spikes, or both (fig 4). Focal spikes were either independent or associated with the generalised paroxysms (preceding, following or interspersed). The same patient could have spikes in multiple locations in the same or a previous EEG. These focal abnormalities had often been interpreted previously as evidence of partial seizures with secondary generalisation.

EEG and precipitating factors

Hyperventilation facilitated the spike-wave discharges in all patients. Photoconvulsive responses were recorded in eight (34.8%) patients (fig 6). One patient (case 7) had absences on rubbing her face, particularly around the nose, was photosensitive (fig 6) and had myoclonic jerks and GTCS on awakening. Another patient (case 16) with clinical and EEG photosensitivity from 12 years of age, developed late-onset absences precipitated by viewing complex patterns (carpets) confirmed by appropriate EEG recordings.

The background EEG was normal except in chronic and heavily medicated patients where

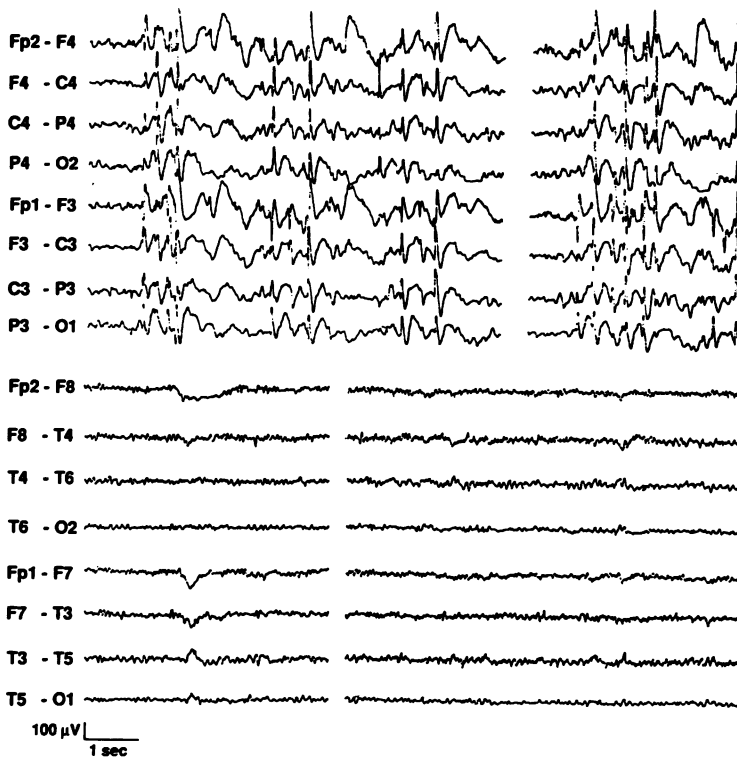


Figure 4 EEG recording of a 40 year old woman (case 4) with juvenile myoclonic epilepsy (JME) (absences at 7 years, myoclonic jerks at 10 years and GTCS at 12 years). Initially diagnosed as suffering from complex partial and secondary generalised seizures, she had one or two GTCS per month and one to three absences per day while on carbamazepine and phenytoin. Absences and myoclonic jerks stopped after carbamazepine was replaced by sodium valproate. She had five nocturnal GTCS in the last two years of follow up. CT of the brain was normal. The EEG (upper trace) showed the typical JME pattern with Ws, discharge fragmentations, inconsistent spike and/or multiple spike-slow wave relation and intradischarge frequency variations. Focal spikes, independently right or more frequently left, were seen either at the onset or within the discharges. The resting EEG (lower trace) had also transients of slow waves localised either on the left or right mid-temporal electrodes.

an excess of diffuse theta activity was found (fig 4).

Syndromic classification of the patients

Table 1 shows the syndromic classification of the 23 patients.

Seven patients (cases 1–7) had juvenile myoclonic epilepsy (JME).^{21–23} All had typical absences, myoclonic jerks on awakening and GTCS (except case 1 who did not have GTCS). Clinical and EEG manifestations of typical absences (figs 4 and 6) were as previously described in adolescent patients with JME.²¹

Eyelid myoclonus with absences^{27–28} was diagnosed in three patients (cases 8–10). Rhythmic eyelid myoclonus was documented by EEG and video-EEG during the absences. Photosensitivity was found in one of the three (case 10). Typical absences varied in duration from 3–12 s. Impairment of consciousness was more severe than in JME but not as intense as in juvenile absence epilepsy (fig 2).

Juvenile absence epilepsy (JAE)^{21–24–29} was the likely diagnosis in four patients (cases 11–14). Two patients (cases 11 and 12) had typical absences with onset at puberty. They also had GTCS and occasional diurnal mild myoclonic jerks. One patient (case 13) had

pyknolepsy from age 12, clinical remission at age 20 and a relapse at age 30 with mild and infrequent absences. He had no other type of seizures. Another (case 14) had onset of intractable pyknoleptic absences at age 8 years which continue to date with the same frequency and severity; she has never had any other type of seizures. Typical absences in these patients showed more severe impairment of consciousness than in any other epileptic syndrome in this study (fig 1).

Late-onset absence status with GTCS³⁰ (case 15), photo- and pattern-sensitive epilepsy³¹ (case 16) and symptomatic late-onset absences³² (case 17, fig 5) were also classified with relative confidence.

The remaining six patients had uncertain syndromic classification (see table 1 and discussion).

Diagnosis on referral and treatment

None of the 23 patients had a syndromic diagnosis on referral. Furthermore, absences were not recognised in 14 patients and in eight of them, the working diagnosis was of complex partial seizures.

Seizures in the majority of patients were not controlled on referral, despite often multiple anti-epileptic drugs. Carbamazepine was the most widely used drug (nine patients) followed by sodium valproate (eight), phenytoin (six), phenobarbitone (four), sulthiame (one), vigabatrin (one), ethosuximide (one). None of the patients were on clonazepam.

The follow up period was short (maximum two years) for quantitative conclusions regarding treatment, mainly with sodium valproate, and slow withdrawal of other anti-epileptic medication. Four patients were free of seizures, a 60–80% reduction of all types of seizure was seen in 10 and there was no change in three patients. Data for the other three of the 20 unselected patients were incomplete.

Discussion

All patients—adults with normal neurological and mental status—had typical absences as defined by the Commission of the ILAE.²⁰ This report refers to patients with overt clinical absences documented by EEG or video-EEG recordings, or both, and not to patients with EEG epileptiform discharges unaccompanied by clinical changes (subclinical, larval or electrical seizures).²²

We have documented that typical absences in adults show a considerable syndrome-related variation in their combined clinical presentation and EEG features. Impairment of consciousness varies from severe to mild and is frequently difficult to document objectively with conventional means. This is in agreement with our previous study on children and adolescents.^{23–24}

Clinically, the absences were perceived by the patients as transient sensations of “momentary lack of concentration”, “flashes of blackouts” which could be misinterpreted as normal sensations or drug-induced in patients who were often overmedicated. We, the clini-

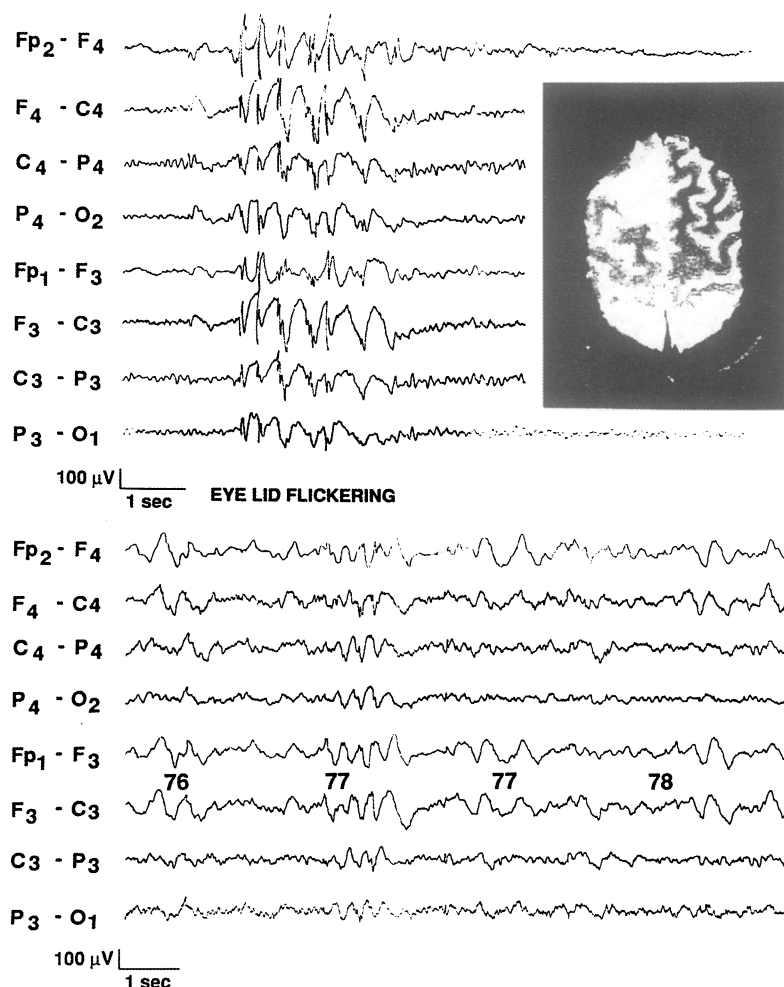


Figure 5 Symptomatic absences of a 30 year old female with late onset absences and GTCS. Her initial EEG (left) demonstrated absences with eyelid flickering. Impairment of breath-counting was documented during brief generalised discharges in subsequent video-EEG recordings (right). CT scan demonstrated a left frontal lesion, probably a low grade glioma.

cians, have been trained to identify absences in their classic form,^{20,21} that is, of a child with transient episodes of loss of consciousness. We are not familiar with absences in an adult whose level of awareness is not severely impaired during the absence ictus. We have demonstrated in this study that absences in adults may escape recognition because they are often mild. This significant observation has been made in only one other clinical report: "... in the patients who continued to have petit mal, the duration of the attacks was usually one second or less, and they were often not noticeable to family, friends, or school-mates".⁶ Extensive studies have demonstrated that the generalised spike-wave discharges described as "subclinical, larval or electrical seizures" are frequently associated with cognitive impairment (see references 22 and 33 for review).

Furthermore, absences may escape clinical detection, even during conventional EEG. Current practice is to test, in an all-or-none manner, the ability of a patient to recall a phrase or number administered during a generalised spike-wave discharge. It has been shown in this study that this is often not adequate. We have frequently reviewed video-EEG of patients with spike-wave discharges in

whom clinical manifestations (such as slowing down of breath-counting, repetition and mistakes, or stuttering) became apparent only after careful video-EEG viewing.

For practical purposes we propose that patients with spike wave paroxysms should be asked to count their breaths during hyperventilation; this may show slowing, discontinuity, errors, repetition and other signs of impairment of consciousness and speech which otherwise would escape recognition with the current practice established in EEG departments.^{23,24} This method is sensitive (involves concentration, memory, recollection of learned experience, expressive speech and other cognitive functions), practical (easy to perform by the patient and easy to evaluate by the observer) and is clinically relevant (reflects impairment of day-life performance).

The frequency of 10% of typical absences found in adults in this study is the highest ever reported.¹⁻¹⁹ Absences were undiagnosed in more than half of our patients on referral, a proportion comparable to that of misdiagnosed absence status despite its long duration (hours or even days) and the associated severe manifestations of confusion and impaired behaviour.³⁰

Underestimation of absences may be even more extensive than in this study; impairment of consciousness is often manifested in patients with series of repetitive myoclonic seizures or myoclonic status, or both, as illustrated by another of our more recent patients (fig 7).

In one third of our patients the absences were misinterpreted as complex partial seizures. Typical absences in adults are brief, lasting only for seconds with, usually, mild impairment of consciousness. It is because of this mild impairment that automatisms were so rare in our patients compared with the frequent automatisms occurring in childhood and juvenile absences.²⁴ Conversely, complex partial seizures are of longer duration (minutes) and are associated with prominent emotional and sensory perceptions of the patients with frequent automatisms.

Despite these differences, the diagnosis of complex partial seizures was maintained, even after EEG data were available, either because the physician was reluctant to change his or her clinical impression because of an EEG report showing "generalised discharges without associated clinical manifestations", or because mild, occasionally persistent, focal EEG abnormalities were misinterpreted as indicating partial seizures. Focal EEG abnormalities have been reported and are accepted in childhood absence epilepsy³³ but are not generally known in other idiopathic generalised epilepsies, such as JME, despite their documentation in this and previous reports.^{23,34,35}

Therapeutic decisions were influenced by the diagnostic problems. Sodium valproate, which is the drug of choice in idiopathic generalised epilepsies³⁶ was prescribed less often than carbamazepine, which is not appropriate treatment for absences or myoclonic epilepsy³⁷ (its effect in GTCS of idiopathic generalised epilepsies has not been clarified).

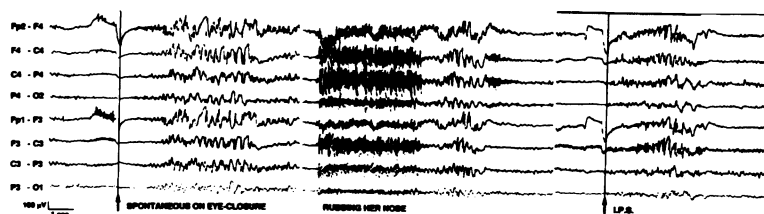


Figure 6 EEG of a 26-year old nurse with JME. Generalised spike and occasional multiple spike and slow wave discharges are recorded on eye-closure and are evoked by somatosensory stimuli (rubbing her nose) and intermittent photic stimulation.

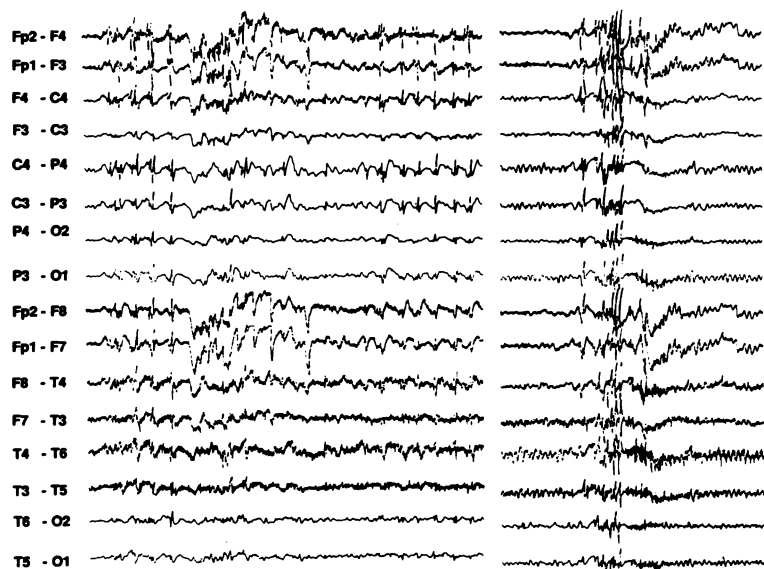


Figure 7 EEG of myoclonic status of a 36-year-old woman with JME while on carbamazepine and phenytoin. Myoclonic jerks and GTCS, but not absences, were elicited on history. Typical EEG features of myoclonic jerks (right) and absences (left) were recorded. In addition, eyelid myoclonus was noted by the technician during this myoclonic-absence status.

Ethosuximide³⁸ and clonazepam³⁹ were rarely used, despite their importance as adjunct therapy in patients with refractory absence syndromes. Our patients showed considerable improvement in frequency of all seizure types (absences, myoclonic jerks and GTCS) on sodium valproate. Longer follow up is needed, however, to confirm these results, as well as the efficacy of ethosuximide and clonazepam as a combined treatment with sodium valproate.

The significant predominance of women in this study is similar to that found in childhood absence²¹ and photosensitive epilepsy³¹ but unlike the male predominance of juvenile absence epilepsy²⁹ and the equal sex^{23 34 35} distribution of JME. It may be attributed to the finding that remission of typical absences is more likely in boys than in girls.¹³

We attempted an epilepsy disease/syndrome diagnosis of the 23 patients with absences, in accordance with the proposal of ILEA.²¹ It is beyond the scope of this report to discuss limitations and drawbacks of this proposal (such as inadequately defined and overlapping syndromes, bias towards criteria supported by one but not other authors, complex and difficult terminology) which are well illustrated in this study. This should not be used as an argument against a syndromic classification of epilepsies. Common syndromes such as JME

are well defined^{21 23} and a confident diagnosis could be made for the majority of the patients reported here. The following discussion aims to promote the use of a medical disease/syndrome diagnosis of patients with seizures because this is probably the only way out of the monoleptic diagnosis "epilepsy" which our patients are often labelled with for life.

Juvenile myoclonic epilepsy with the triad of age-related idiopathic generalised seizures, myoclonic jerks on awakening and the characteristic clinical and EEG manifestations of typical absences,^{23 24} was easy to recognise in seven of our 23 patients. This study confirms our previous reports^{23 24} that typical absences in JME have distinct combined clinical and EEG features.

Ictal eyelid myoclonus with absence seizures was consistently recorded in three patients; it was also occasionally seen in one patient with symptomatic absences (case 17) and may also be observed during myoclonic status (fig 7). Only one of the three patients would meet the criteria of the syndrome described by Jeavons,²⁷ that is, photosensitive patients with absences that are resistant to treatment and show marked jerking of the eyelids associated with spike wave discharges on eye-closure. Although not representing a monosymptomatic criterion for this classification, ictal eyelid myoclonus with absences may be used as a basis for a syndrome as yet ill-defined, not recognised in the ILAE classification²¹ and probably wider²⁸ than that involving photosensitive patients only.

Four patients were classified as having juvenile absence epilepsy^{20 23 25} on the basis of age at onset, combined ictal clinical and EEG manifestations²⁴, frequency of seizures and exclusion criteria (lack of myoclonic jerks on awakening, lack of facial or limb myoclonus during the absences or early-onset GTCS).

Juvenile and childhood absence epilepsy are sometimes difficult to differentiate.^{20 25} The ILAE proposal²¹ uses criteria of age at onset, frequency of the seizures and the retropulsive movements of the eyes and head (a symptom consistently found in eyelid myoclonus with absences but rarely in other forms of childhood absences). Combination of ictal clinical and EEG features of absences together with some of the ILEA criteria may provide a more satisfactory syndromic classification. Exclusion criteria, for example myoclonic jerks on awakening and eyelid myoclonus, may be as important as inclusion criteria.

Classification of three patients with late onset absence status³⁰ (case 15), photosensitive and pattern sensitive epilepsy with absences³¹ (case 16) and symptomatic epilepsy with typical absences³² (case 17) was straightforward.

The remaining six patients could not be classified with confidence. Case 18 had absences with EEG but without clinical documentation, as early as five years of age. JME was a possible diagnosis but myoclonic jerks were mild and did not show the characteristic clustering on awakening. Juvenile absence epilepsy was also likely, in view of age at onset of

clinical manifestations of absences, associated symptoms and other seizure-types. Childhood absence epilepsy was unlikely, in view of mild impairment of consciousness.²⁴

Case 19 had the seizure triad of JME but myoclonic jerks appeared very late (50 years of age) and did not cluster on awakening. Age at onset of absences and absence status with mild myoclonic jerks could not be determined.

Cases 20 and 21 had severe absences with consistent perioral myoclonus and late onset GTCS. Absence status with perioral myoclonus occurred in case 21. Childhood (case 21) or juvenile (case 22) absence epilepsy would be the diagnosis dictated by the present ILAE classification.²¹ Consistent perioral myoclonus in absences may, like absences with eyelid myoclonus, be a feature of a separate epileptic syndrome which is resistant to treatment and persists in adult life. This is supported by another three patients with absences and perioral myoclonus that we have studied recently. Perioral myoclonus consists of rhythmic profusion of the lips, such as, the mouthing movement of goldfish.

Case 22 may be a case of late-onset absence status, absences and GTCS. The remaining patient (case 23) with late-onset GTCS, was unaware of absences from which she may have suffered for years. A mild form of juvenile absence epilepsy was a possible diagnosis.

Irrespective of arguments for or against the proposed classification of the patients, the clinical and EEG manifestations described may help in the recognition, differential diagnosis, prognosis and treatment of adult absence syndromes, which are not unusual. Furthermore, this study may encourage revision of the classification of epileptic syndromes with typical absences based on prospective clinical and video-EEG studies.

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